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Research Letter

Gastrointestinal stromal tumor mimicking ovarian malignancy in a woman with type I neurofibromatosis

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A gastrointestinal stromal tumor (GIST) is a specific mesenchymal tumor of the gastrointestinal tract [1]. It is believed to arise from the interstitial cells of Cajal [2]. A GIST can originate anywhere in or near the gastrointestinal tract such as the stomach, bowel, rectovaginal septum, and uterus [2–6]. It may also present as a pelvic mass that mimics a gynecologic neoplasm [3]. To date, only a few cases of GIST mimicking gynecologic tumors have been reported [2–6]. Most gynecologists are likely to be unfamiliar with the clinical features and outcome of a pelvic GIST. In this paper, we describe the clinical features, treatment, and outcome of a pelvic GIST in a middle-aged woman who was initially diagnosed with a possible ovarian malignancy.

A 54-year-old woman, gravida 4, para 2, with type I neurofibromatosis presented at our hospital with abdominal distension that deteriorated progressively in the months before her visit. Ultrasonography showed a solid right adnexal mass and minimal ascites. Computed tomography (CT) confirmed a 14-cm pelvic mass containing cystic and solid components (Figure 1A), dilated bowel loops, and left hydronephrosis. Laboratory data showed a low level of serum hemoglobin (7 g/dL) and elevated level of serum cancer antigen (CA) 125 (363.6 U/mL). Preoperative colonoscopy and gastroscopy did not show any intraluminal mass and other tumor markers; in particular, carcinoembryonic antigen and cancer antigen 19-9 were within normal limits. We preoperatively suspected an ovarian malignancy.

Exploratory laparotomy was performed and a 14-cm necrotic and semisolid tumor was found. It was densely adhered to the uterine fundus and ileum. Frozen section pathological examination revealed that the mass was a sarcoma. Total abdominal hysterectomy, bilateral salpingo-oophorectomy, bilateral pelvic lymph node dissection, and partial omentectomy were then performed with segmental resection and reanastomosis of the ileum because of tumor-related segmental stenosis. No residual tumor remained after surgery.

The final histopathologic examination showed that the tumor was a malignant GIST (Figures 1B and 1C). The mitotic index of the tumor was 5–10 per 50 high power fields. The tumor cells stained positive for CD117 (diffuse), S-100 (focal), and vimentin, and stained negative for actin and desmin. The patient received imatinib (400 mg daily) during a 3.5-year follow-up period, and her postoperative course was uneventful. The CA125 levels declined after surgery (51.12 U/mL at 1 month postsurgery, 11.88 U/mL at 2 years postsurgery, and 10.37 U/mL at 3 years postsurgery). At the 3.5-year follow up, there was no evidence of recurrent GIST.

In this patient, our initial preoperative diagnosis of a possible ovarian malignancy was incorrect. In general, it is often difficult preoperatively to diagnose GIST correctly because it is a rare mesenchymal tumor that can occur anywhere in or near the gastrointestinal tract. Imaging modalities such as CT, magnetic resonance imaging, and fluorine-18-fluorodeoxyglucose positron emission tomography are useful diagnostic and treatment evaluation tools for a suspected GIST. On CT, a large well-circumscribed densely enhancing mass with an exophytic growth pattern or ulceration suggests a diagnosis of GIST. However, it may not be possible to differentiate GIST from other pelvic neoplasms using CT alone. CT nonetheless can also be useful in tracing the tumor pedicle and detecting hepatic or peritoneal metastatic disease [7]. The combination of positron emission tomography and CT may be optimal for the precise delineation of the tumor and assessment of the patient's response to therapy [7].

Complete resection remains the most effective treatment for GIST [8], whereas retroperitoneal lymph node dissection is recommended in cases of ovarian cancer. In GIST, metastasis to

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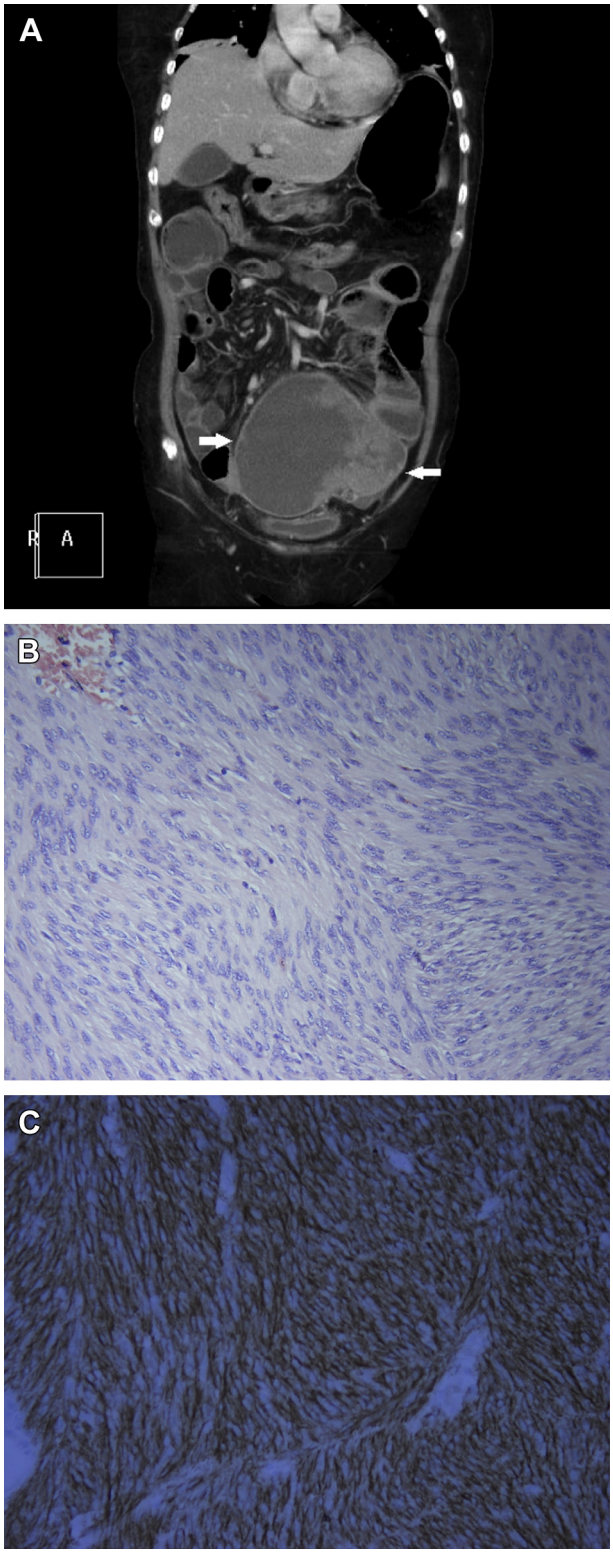

Figure 1. (A) The coronal computed tomography scan of the pelvis shows a dilated loop of the jejunum with uneven wall thickening and an adjacent large, extramural, necrotic pelvic mass (arrows). Histopathological examination shows (B) intersecting fascicles of spindle tumor cells [hematoxylin and eosin (H&E) staining; magnification, 200×] and (C) strongly positive immunohistochemical staining for CD 117 in the spindle tumor cells (H&E staining; magnification, 200×).

regional lymph nodes is not typical and lymphadenectomy is not routinely required [8]. Debulking surgery such as pelvic lymph node dissection and partial omentectomy was performed in our patient, although lymphadenectomy and partial omentectomy can be omitted if the pathologic examination of a frozen tumor section leads to a diagnosis of GIST.

A coordinated surgical approach that includes a gynecologist and general surgeon should be considered in cases of pelvic GIST with intestinal involvement. Imatinib remains the mainstay adjuvant therapy for GIST after surgery [8]. In general, the outcome of GIST is good. At 12 months posttreatment initiation, 71% of patients reportedly show no progression of disease and the overall survival is 86%, even in patients with advanced GIST [8]. Our patient had type I neurofibromatosis. Gastrointestinal stromal tumors are approximately 150 times more common in patients with type I neurofibromatosis than in the general population [9].

In conclusion, a pelvic GIST may mimic an ovarian neoplasm but it requires a different surgical approach. A GIST is a rare disorder and it should be included in the differential diagnosis of a pelvic mass—especially in women with type I neurofibromatosis.

Conflicts of interest

The authors have no conflicts of interest relevant to this article.

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